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Surgical Removal of Dysplastic Aortic Valve: A Case Report

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Cardiac tumors are infrequent, comprising 0.1% to 0.2% of all cardiac diseases[1]. In almost all of these cases, the tumors are attached to the free wall or the septum, and tumors attached to valves are very rare. One of our patients, a male infant, had a tumor attached to the wall of the left ventricular outflow tract demonstrated by echocardiography and cardiac catheterization before operation. At operation the tumor was found to have originated from the right cusp of the aortic valve. Microscopic examination revealed a dysplastic aortic valve.

Case Report

The patient was born at 38 weeks of gestation with a birth weight of 2850 gm. No cyanosis or asphyxia was noted at birth. At the age of 4 months, cardiac murmur and tachycardia were noted, and at the age of 6 months, he was hospitalized because of cardiac failure. At the age of 9 months, cardiac catheterization revealed a mobile tumor in the left ventricular outflow tract. The tumor was considered to have caused aortic stenosis, mitral valve insufficiency and pulmonary hypertension. He was referred to our service for open heart surgery. On admission, he weighed 6.2 kg (normal 9.0 kg) and he had tachypnea (62/min.). Physical examination showed a grade 4/6 high pitched pansystolic murmur at 2RIS and a grade 2/6 pansystolic murmur at the apex. The liver was palpable 4 cm below the right costal margin. The chest X-ray showed cardiomegaly with a CTR of 74%. An electrocardiogram demonstrated left ventricular hypertrophy and bilateral atrial hypertrophy. Cardiac catheterization confirmed pulmonary hypertension (mPA 70 mmHg/15 mmHg), and a gradient of 95 mmHg was recorded between the aorta and the left ventricular outflow tract. A pulmonary angiogram showed a negative shadow that moved across the aortic valve. (Fig. 1) An echocardiogram disclosed a mobile mass in the aorta during systole and within the left ventricue dulring diastole. From these findings, a diagnosis was made of aortics tenosis, mitral regurgitation, and pulmonary hypertension caused by a tumor attached to the wall of the left ventricular outflow tract.

Key words: Cardiac tumor, Dysplastic aortic valve.
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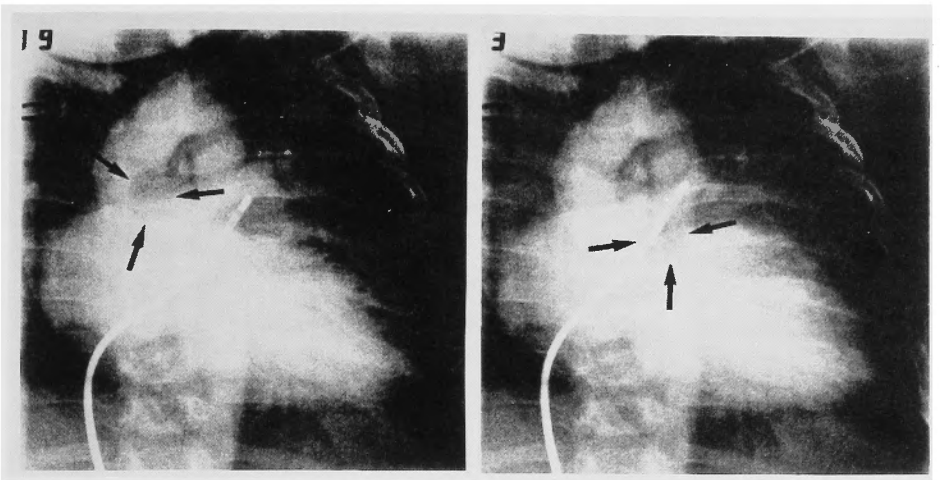

Fig. 1 PA graphy. Arrows show negative shadow of tumor.
Right: Systolic phase. Left: Diastolic phase.

Operative Findings

Open heart surgery was performed under cardiopulmonary bypass and cold potassium cardioplegia for myocardial protection. The aortic root was opened, and a "cauliflower-like" tumor came into view (Fig. 2). The tumor arose from the right aortic cusp, not from the wall of the left ventricular outflow tract. Since the boundary between the cusp and the tumor was not clear, it was impossible to excise only the tumor. Aortic valve replacement was performed. The diameter of the aortic annulus was 14 mm. To insert a 17 mm Björk-Shiley mechanical prosthesis, we expanded the aortic annulus by the methods of Nicks[2] and of Blank[3].

Postoperative Course

Low cardiac output syndrome and respiratory insufficiency continued for 6 days after the operation, at which time he was weaned from the respirator, but from the 9th day the patient exhibited respiratory insufficiency, which necessitated reintubation and respiratory support. On the 12th day, the tube was removed. However, he developed intermittent fever and was given antibiotic therapy. Unfortunately, the patient died of sepsis on the 16th day after operation. Autopsy was not permitted.

Pathology of the Tumor

Macroscopically, the tumor was 4.5×1.5 cm and weighed 12 g. It had a "cauliflower-like" shape and contained a cyst. Histologically, the tumor had fusiform cells with myxomatous stroma. It resembled a myxoma, but it lacked vessels and elastic fibers. No hemorrhage, hemosiderin, or invasion of inflammatory cells was noted. The specimen was sent to the AFIP (Armed Forces Institute of Pathology, washington D.C., U.S.A.), where it was diagnosed as a "dysplastic cardiac valve."

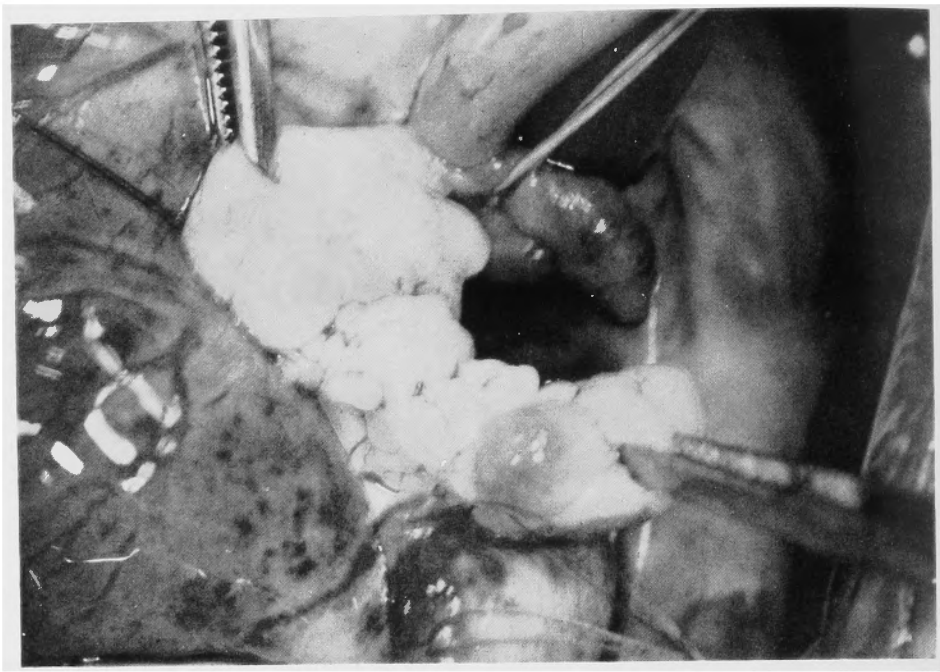

Fig. 2 A "cauliflower-like" tumor came out from the incision of the aortic root.

Discussion

A primary cardiac tumor is a very rare anomaly of the heart. Its incidence was 0.05% in a series of 8,550 autopsies performed at the Mayo Clinic. Among cardiac tumors, 80% are benign[4], 50% of the benign tumors are myoxmas and 20% rhabdomyomas. Most of the malignant tumors[5] are sarcomas or metastatic tumors[6]. Benign cardiac tumors are generally attached to the endocardium, intramyocardium, or pericardium. Of the intracardiac tumors, the majority are myxomas, but these are very infrequent in childhood. Most of the intramyocardial tumors are rhabdomyomas, which are seen even in childhood. Half of the epicardial tumors are sarcomas or mesotheliomas in adults. Reports of tumors that arise from valves are very few, but BUTTERWORTH[7] described a case of papilloma. Our patient was first diagnosed as having congenital aortic valve stenosis because of progressive heart failure and cardiac murmur. Further examination by echocardiography, cardiac catheterization, and pulmonary angiography revealed a tumor arising in the left ventricular outlow tract. We suspected a myxoma of the left ventricle which moved through the aortic valve rather than a tumor originating from the aortic cusp on the basis of echocardiographic and pulmonary angiographic findings. However, NADAS reported that myxoma occurs infrequently in childhood and originates in the left atrium in more than 75% of the cases and is rarely found in the left ventricle. Since YOUNG[8] described the first pediatric patient with myxoma, only 20 additional cases have been reported. In regard to tumors originating from a valve, ANDERSON described a case of congenital papillary tumor of the tricuspid valve. WOLD reported that among 68 cases of myoma, the tumor originated from

a valve in 7 cases but from the aortic valve in only one case. Regardless of whether the tumor originates from a valve or not, it is important to differentiate myxoma from dysplastic valve histologically. This tumor occurred in childhood and did not show vessels, hemosiderin, or invasion by inflammatory cells, which are characteristics of myxomas. HYAMS[9] included "dysplastic valve" in his list of incomplete differentiations of cardiac valves: myxomatosis, myxoma, mucinous fetal endocardiosis, dysplasia of leaflets, fibromyxoma, fibromyxomatous hyperplasia, congenital myxoma, myxoid dysplasia, and hamartoma. ABBOT[10] also described dysplastic cardiac valves occurring when the fetal valve develops abnormally. HYAMS reported that among 197 cases of tumors originating from the aortic valve 55 (28%) showed incomplete differentiation of the valve, and almost all of these had thickened and deformed valves; in a few cases, the tumors were in the ventricular cavity or aortic lumen. Echocardiography and computed tomography are sensitive noninvasive methods for detecting cardiac tumors, especially cross-sectional echocardiography[11,12]. Angiocardiography is used for definitive diagnosis in almost all cases. STEINER[13] emphasized that only pulmonary arteriography should be performed to prevent embolism by displacement of the tumor by the catheter. Generally, if the tumor originates from the internal ventricular wall and has a stalk, only the tumor should be removed. If there is no clear bundary between the tumor and the cusp, as in our patient, it is difficult to dissect it. We thank Dr. W. R. COWAN and Dr. M. RABINOVITZ for help with the histological diagnosis.

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和文抄録

Dysplastic aortic valve の1例

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心臓腫瘍は稀な疾患である．その中でも弁膜より発生する腫瘍は極めて稀であり，我々は大動脈弁のDYSPLASTIC VALVE の一例を経験したので報告する．患児は11ケ月男児で，主訴は発育障害である．術前心エコー及び心臓カテーテル検査で左室流出路壁より発生した腫瘍による大動脈弁狭窄症，僧房弁閉鎖不全症および肺高血圧症と診断された．心筋保護のもとに開心術を行い大動脈弁無冠動脈尖より発生した腫瘍であることが確認された．組織学的には粘液腫に類似するが AFIP の診断によれば DYSPLASTIC CARDIAC VALVE であり NEOPLASMA でなくDYSPLASIAであることが判明した．大動脈弁より発生する腫瘍は極めて稀であり，現在までPAPILOMAの一例のみが報告されているに過ぎず，本邦では最初の報告である．弁置換術を行ったが，術後16日目に，敗血症で死亡した．